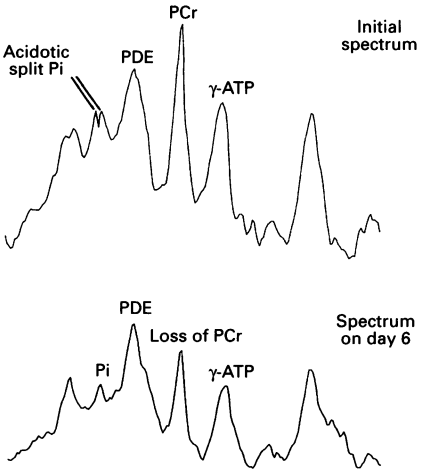


*Phosphorus spectra from the right hemisphere. The initial spectrum on day 3 showed a split inorganic phosphate (Pi) peak, suggesting that there was tissue of normal pH and acidotic tissue with pH 6·35. The second spectrum, of the same region on day 6, showed a loss of adenosine triphosphate (ATP) and phosphocreatine (PCr), associated with clinical deterioration. PDE = phosphodiester peak.*

relative to the cerebellum 0·58 (0·06), significantly ($P < 0·05$) less than a symmetric region of interest from the left frontal lobe, 0·87 (0·04)).

Despite these SPECT and spectroscopy changes, there was no evidence of vasospasm. Daily transcranial Doppler studies (using a Scimed PC Dop machine) demonstrated middle cerebral artery velocities within the normal range (right mean velocity (SD) 43 (4·3) cm/s, left 35 (5·6)). Angiography, performed for clinical evaluation on day 7, showed a middle cerebral artery aneurysm, and confirmed the absence of vasospasm.

Subsequently the patient recovered, the aneurysm being clipped on day 11. Peroperatively she re-bled, developing a left hemiplegia apparent immediately after the operation. Postoperative recovery was further complicated by vasospasm detected with transcranial Doppler. This resolved, and the SPECT was repeated on day 25. Perfusion had increased in the right frontal lobe, which was significantly ($P < 0·05$) hyperaemic (0·98 (0·08) compared with 0·82 (0·06) contralaterally). There was, however, an area with persistently low counts (0·63 (0·06)) in keeping with an established infarct.

The clinical deterioration was typical of delayed ischaemia, six days after the presenting haemorrhage. The SPECT showed hypoperfusion, and spectroscopy a sequence of events reflecting ischaemia with lactic acidosis progressing to infarction. Repeat CT excluded hydrocephalus and rebleeding as causes of the deterioration, and in the absence of any metabolic derangement, a diagnosis of clinical vasospasm might be made. The striking finding was the absence of vasospasm either angiographically, or with serial transcranial Doppler studies. Angiography remains the "gold standard" for diagnosing vasospasm. The angiogram, first SPECT, and second spectroscopy examination were all performed within 24 hours of the clinical deterioration. As vasospasm persists for days, if it was the cause of the deterioration we would have expected to see it. Angiography and transcranial Doppler, however, examine the large and medium sized intracranial arteries, whereas SPECT assesses perfusion through the microcirculation. The normal angiography and transcranial Doppler examinations, but abnormal SPECT, suggest that there may be relevant changes in small vessels beyond the resolution of the angiogram.

This is similar to a case described by Soucy *et al.*[4] Hypoperfusion on SPECT was not associated with vasospasm on angiography performed two days before, or five days later, raising the possibility of "subradiological vasospasm". There is also support for the existence of small vessel disease from postmortem studies, changes in the small arteries and capillaries being noted, but presumed to be the result of ischaemia, rather than the cause of it.[5]

The changes in this patient must have started by day 3 to account for the acidosis seen spectroscopically. The improvement in perfusion by day 25 (despite additional operative trauma and postoperative vasospasm), suggests that any change can reverse in three weeks. This is similar to the time course of angiographic vasospasm. Striking radiological changes in the large vessels may have drawn attention away from the possibility of coexistent changes in the small vessels. Such changes may be as important, or in some patients like this, more important than large vessel changes. This may explain discrepancies between angiographic vasospasm and cerebral ischaemia.

This concept of small vessel changes does not render angiographic vasospasm unimportant. If small vessel resistance increases, any vasospasm affecting large vessels may compromise the circulation much more than if the small vessels are unaffected. Findings of impaired cerebrovascular reactivity accounting for discrepancies between angiographic vasospasm and cerebral ischaemia may also be explained. These adaptive responses occur primarily at a level of the small vessels, so measuring changes in cerebrovascular reactivity may be a functional assessment of the small vessel changes detected in these imaging studies.

In summary, the vascular changes complicating subarachnoid haemorrhage may be more extensive than is appreciated angiographically, extending to the small vessels. If such changes coexist with angiographic vasospasm, this could explain why angiographic changes correlate poorly with clinical ischaemia.

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## Neurosyphilis presenting with dissociative symptoms

A 62 year old man was admitted as an emergency to a medical ward with a five day history of expressive dysphasia. The onset was acute and the dysphasia variable, becoming more pronounced when he was anxious. He had a 20 year history of insulin dependent diabetes, but there was nothing else of note. There was no psychiatric history. His wife was also in hospital; she had been diagnosed as having probable Creutzfeldt-Jakob disease three weeks before the onset of his difficulties. Her deterioration after initial presentation had been rapid, and she also had pronounced expressive dysphasia in the late stages of her illness. Before admission, the man had an episode of expressive dysphasia after visiting his wife in hospital and another while on the ward visiting her. His wife's resuscitation status and the possibility of a postmortem had been discussed with the patient within 24 hours of the onset of his speech problems.

According to a mental state examination carried out after admission, he was very anxious and agitated; the agitation and dysphasia prevented cognitive function testing at this point. Physical and neurological examination were normal. Investigations at this time showed that full blood count, plasma viscosity, urea and electrocytes, liver function tests, thyroxine, and thyroid stimulating hormone were all within normal limits; plasma glucose was raised at 14·2 mmol/litre. A chest radiograph and ECG were normal, as was a carotid scan. In view of the circumstances of the presentation, a provisional diagnosis of a dissociative disorder was made and a psychiatric opinion sought.

Ten days after admission, he became increasingly agitated, emotionally labile, and tearful, and his speech was so slurred that cognitive assessment was still not possible. In view of this deterioration, he was transferred to an acute general psychiatric ward. His wife died eight days later. He seemed to take this news well, becoming less agitated and dysphasic. Cognitive function testing at this point showed significant impairment (mini mental state examination = 18/30). Brain CT showed advanced cerebral atrophy with no focal abnormalities, and there was excessive slow wave activity in the left temporal and anterior regions on the EEG. Autoantibody screening was normal. In view of these findings, he was trans-

ferred to an organic psychiatric unit for further investigation. The dysphasia resolved, but he became increasingly perseverative in his speech and actions, confabulating frequently, and his behaviour was socially disinhibited. He began to express grandiose ideas. Neuropsychological assessment showed pronounced frontal lobe impairment.

At this point, syphilis serology showed venereal disease research laboratory test (VDRL) positive (1:32), TPHA positive (1:256), and fluorescent treponemal antibody (FTA) positive + + + +. Further investigation of CSF showed VDRL positive (1:4), TPHA positive (1:1024), and FTA positive + + + +. The isoelectric focusing pattern to detect oligoclonal IgG was negative for serum and positive for CSF. A diagnosis of neurosyphilis was made, and oral doxycycline (100 mg thrice daily) was commenced.

His wife did not undergo a postmortem. In view of the patient's diagnosis, reexamination of his wife's stored serum was carried out. A repeat test for syphilis in serum was negative. As part of the investigation of her illness, CSF had been examined, showing normal cells and proteins. Tests for syphilis were not performed on CSF.

The source of the patient's primary infection is unknown. He had been happily married for 40 years. Of possible relevance in his employment history was the fact that he had served in the army for two years in Germany as a transport driver.

This case is remarkable for the apparent coincidence of two unusual causes of dementia in a married couple. There is a possibility that the patient's wife might also have had neurosyphilis; this disorder has been reported as presenting as Creutzfeldt-Jakob disease.[1] Her negative serum VDRL and normal CSF cells and protein make this unlikely, but in the absence of postmortem confirmation of the diagnosis or antemortem CSF examination it is not possible to be certain. Examination of CSF to exclude neurosyphilis should always be carried out in patients with possible Creutzfeldt-Jakob disease.

This case is also a reminder that hysterical dissociation is a very unsafe diagnosis to make in older adults.[2] Even genuinely dissociative symptoms in elderly patients are usually indicative of an underlying organic cerebral disorder, and this should always be investigated. The initial diagnosis of a dissociative disorder in this patient was based on the circumstances surrounding the onset, the absence of focal neurological signs, and the presentation with symptoms which mimicked his wife's. The cognitive impairment only became apparent as the dysphasia and agitation resolved.

The dramatic onset of the patient's symptoms in the context of his wife's terminal illness is interesting. It was commonly reported in the pretreponemal literature that the onset of general paralysis of the insane could be precipitated by "mental shocks" such as bereavement and illness in the family,[3 4] but this clinical finding has never been systematically studied. The acute presentation of people with cognitive deficits is not uncommon when a spouse falls ill or dies. This is usually because the sudden departure of the carer discloses the impairment, but sometimes the stress of the event causes a significant decompensation, as seems to have occurred in this case.

Another lesson to be drawn from this patient is that neurosyphilis is not a historical curiosity, but something that clinicians need to keep in mind particularly when investigating elderly patients. There is evidence that neurosyphilis is becoming clinically less typical,[5-7] so serological investigation is all the more important if the diagnosis is not to be missed. Routine screening of all elderly patients is currently out of favour, although still recommended for those with an organic illness.[8 9] This patient, together with others in whom neurosyphilis has presented as a functional disorder, does raise the question of whether these patients should be screened also, particularly those with an atypical illness that is unresponsive to treatment.[9]

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## Alternating paroxysmal dystonia and hemiplegia in childhood as a symptom of basal ganglia disease

We report a 13 year old girl with an unusual clinical presentation of longstanding progressive cerebellar and pyramidal disorder, episodes of alternating paroxysmal dystonia lasting up to an hour, and hemiparesis of the involved side or sides, reminiscent of alternating hemiplegia of childhood. Magnetic resonance imaging showed hypointensity of the basal ganglia, particularly the globus pallidus, similar to that seen in Hallervorden-Spatz disease, raising the possibility that this may represent an atypical form of the condition.

The patient, now aged 13, was born at term after a normal pregnancy and delivery, to non-consanguineous parents: there were no perinatal problems. At two months of age she was noted to be hypotonic. She had delayed motor milestones, sitting at nine months and walking at 22 months, and was said to be "always clumsy". Tremor and ataxia were noted at two years and have been slowly progressive, particularly since the age of eight: by 12 years she needed to use a frame. Examination showed her to have pyramidal signs in all four limbs as well as considerable titubation and cerebellar signs.

At the age of 10 she developed episodes of painful dystonic posturing of the arm and leg, usually on the right but sometimes on the left, associated with hemiparesis. These initially occurred during sleep and were preceded by a cry. They were often associated with a contralateral headache. They lasted 15 to 45 minutes, and ended abruptly, and she could have several in a month, sometimes more than one a day. They were at first helped by carbamazepine but later recurred, during waking as well as in sleep. After withdrawal of the carbamazepine she developed distressing bilateral attacks that were also associated with drooling and difficulty in breathing: these settled after reintroduction of the medication. She did not respond to benzodiazepines. Flunarizine likewise produced no benefit.

Blood tests including lactate, ammonia, thyroid function, cholesterol, triglycerides, caeruloplasmin, copper, $\alpha$-foetoprotein, renal function tests, calcium, magnesium, arylsulphatase A, hexosaminidase, plasma very long chain fatty acids; renal function tests and urinary oligosaccacharides were normal. Examination of CSF, including assay for lactate and amino acids, was normal, and there were no oligoclonal bands. Measles and rubella antibodies were not detected. An ECG was normal, EEG was mildly dysrhythmic, and visual and auditory evoked responses were both delayed. Nerve conduction studies and nerve and muscle biopsies were normal. Cytochrome oxidase and pyruvate dehydrogenase activity were normal. Repeated CT was unremarkable, but MRI showed notable symmetric low intensies in the basal ganglia, particularly in the globus pallidus, but also the putamen, red nucleus, substantia nigra, caudate nucleus, and thalamic pulvinar. There were also changes in the white matter signals in the region of the U fibres (figure).

Our patient presents an unusual clinical picture, in which the development of a paroxysmal alternating or sometimes bilateral dystonia associated with weakness supervened on the background of a progressive neurological disorder involving cerebellar, pyramidal, and extrapyramidal systems, the aetiology of which remains unclear despite extensive investigation. At their onset, the possibility that these episodes represented a seizure disorder was considered, but this has not been confirmed either by the clinical course or by EEG videomonitoring.

The attacks were not precipitated by movement, nor was there any family history. They thus differed from the familial paroxysmal choreoathetosis described by Mount and Reback[1] and from paroxysmal kinesigenic choreoathetosis,[2] which may be familial or sporadic. In each of these conditions alternating and bilateral attacks have been described. In paroxysmal kinesigenic choreoathetosis the attacks also tend to be much shorter than those of our patient, unlike familial paroxysmal choreoathetosis, in which they may last several hours. The occurrence of hemiparesis in association